P9

소아성류마티스관절염 환자에 동반된 이차성 갑상선, 신아밀로이드증

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Secondary Thyroidal and Renal Amyloidosis in a Patient with Juvenile Rheumatoid Arthritis

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A 15-year old Korean girl presented with foamy urine referred to our department of nephrology. She had JRA for 10 years, was on medication and surgery. She had been intermittently taking non-steroidal anti-inflammatory drugs and hydroxychloroquine. She had no past medical history. On physical examination, she had goiter and didn't have endophthalmos. Her lung sounds were clear, the abdomen was not distended and the abdominal bruit was not audible. We could find pretibial pitting edema of 2+ of both lower extremities. Her urine test was as follows: SG (1.015), pH (6.5) protein (3+), blood (+), 24 hours urine protein quantification (5,207 mg/day). Blood tests showed as follows: hemoglobin (10.1 mg/dL), WBC (5,960/m^3), platelet (34,7000/m^3), urea (15.9 mg/dL), creatinine (1.0 mg/dL), sodium (140.6 mEq/L), potassium (4.9 mEq/L), calcium (8.8 mg/dL), phosphorus (4.8 mg/dL), glucose (91 mg/dL), protein (5.8 g/dL), albumin (2.5 g/dL). At that time, a thyroid function test (TFT) revealed hypothyroidism. The results were Thyroid stimulating hormone (TSH) (100.00 mIU/L), Free T4 (1.44 ng/dL), T3 (1.73 pg/mL). The titer of TSH receptor antibodies was 87.20 U/L. She was started on levothyroxine therapy. Renal biopsy was performed after 2 months because she could not take the posture of prone position. Renal ultrasonography showed increased echogenecity on renal cortex and no hydronephrosis. A renal biopsy revealed heavy depositions of segmental eosinophilic homogeneous materials in the glomeruli, interstitium, and vascular walls that were clearly visible under light microscopy. And heavy depositions of non-branching amyloid fibrils in the glomeruli were evident. under EM.There were not remarkable findings.

Congo-red stain showed apple green birefringence of glomeruli, vascular wall and tubulointerstitial region in light microscopy. And immunohistochemical analysis showed strong immune-staining of SAA in the same areas. In addition, thyroid biopsy also revealed deposition of amyloid fibrils.

Treatment was initiated with oral prednisolone of 15 mg and cyclophosphamide of 75 mg per day. The treatment hadmaintained for one year. Cyclophosphamide was discontinued due to thrombocytopenia and azathioprine was replaced. After 8 months of using cyclosporine, there was eosinophilia that we stopped using it. 3 years of diagnosed AA amyloidosis, she has been undergone maintenance hemodialysis therapy as ESRD and has been in a good condition.

Key Words: 아밀로이도시스, 소아성류마토이드관절염, 스테로이드
Amyloidosis, Juvenile rheumatoid arthritis, Prednisolone